Possible significance of juvenile oral venous angioma as marker of intracerebral vascular lesion

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This 10-year-old child suffered a hemorrhage into the right parietal lobe, the result of a ruptured arteriovenous angioma. From birth, the boy had a venous angioma of the mucous membrane of the cheek, lower lip, and hypoglossal area on the right side. The coexistence of these two vascular defects is most unusual, and venous angioma in early life may suggest the presence of cerebral angioma.

KEY WORDS - venous angioma □ intracerebral arteriovenous malformation □ mucous membrane □ tongue

Cerebral angiomas may coexist with angiomas of the internal organs, skin, or mucous membranes. Among the best known syndromes characterized by lesions involving multiple tissues are those from the group of phacomatoses: Sturge-Weber syndrome, von Hippel-Lindau disease, and Ullmann systematic angiomatosis. Telangiectasia of the skin and mucous membranes in Rendu-Osler disease may coexist with cerebral angiomas. Branch, et al., reported the only case involving both a venous angioma of the sublingual region and a venous angioma of the brain. Those authors suggested that the appearance of an angioma of the sublingual region in early youth may signal the existence of a cerebral angioma. Taking this into consideration, we would like to present the case of a patient with an angioma of the mouth who was found in later years to harbor an arteriovenous angioma in the brain. This seems to support the suggestion of Branch, et al., regarding the significance of oral venous angioma in early youth.

Case Report

This 10-year-old boy had suffered since birth from an angioma of the mucous membrane of the cheek, lower lip, and hypoglossal area on the right side. When he was 18 months old, this disorder was treated by freezing, with no great effect. Attempts at treatment by radiotherapy were also unsuccessful. At the age of 4 years, the lesion in the hypoglossal area was removed surgically; the histological diagnosis was venous angioma. The child's parents were healthy, never having been seriously ill. An elder sister has, from birth, had an angioma on the skin of the left hip measuring 3 × 4 cm.

Our patient developed normally until August, 1981, when he was 10 years old, although his performance at school was below average. At that time, he suddenly complained of severe headache and vomited. A moment later he lost consciousness for several minutes. On regaining consciousness he complained of severe headache and was restless. In the course of several hours left-sided weakness developed which increased to hemiplegia. He was admitted on the same day to the Clinic of Neurosurgery in Katowice. On admission he was conscious and oriented as to place, time, and situation. The optic fundi were normal. There were no signs of meningeal irritation. He had paralysis of the left facial nerve of central type, flaccid paralysis of the limbs on the left side with very lively reflexes, and a positive Babinski sign. There was a venous angioma on the mucous membrane of the cheek, lower lip, and the right hypoglossal region (Fig. 1).

Computerized tomography revealed an intracranial hematoma in the right parietal lobe. Angiography of the right carotid artery showed an arteriovenous angioma in the right parietal lobe fed by the parietal branch of the middle cerebral artery and by end branches of the pericallosal artery (Fig. 2).

The arteriovenous angioma was surgically removed, complete with the intracerebral hematoma. Histological
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examination showed an arteriovenous angioma (Fig. 3). The postoperative course was uncomplicated and the child was sent home after 3 weeks. At the time of discharge from the hospital he was walking unaided and exhibited only discrete left-sided paralysis.

Discussion

Birthmarks on the face, trunk, and limbs lead to suspicion of vascular malformations in the brain. Is the same role played by oral venous angioma? Such an angioma is often seen in later years but is a rarity in youth. In later life, oral angioma may coexist with angioma of the intestines, which may cause hemorrhage from the digestive tract. The 19-year-old boy reported by Branch, et al., had a venous angioma of the hypoglossal region together with a venous angioma of the ipsilateral hemisphere of the brain. Cerebral angiomas most frequently make their presence known between 10 and 30 years of age.

In our patient the oral venous angioma was present from birth. Hemorrhage from the cerebral arteriove-

FIG. 1. Photograph of the buccal, labial, and sublingual venous angioma.

FIG. 2. Right carotid angiogram, lateral view, demonstrating the arteriovenous malformation of the parietal lobe and its feeding arteries.

FIG. 3. Photomicrographs of the excised mass. **Left:** A large number of vessels with irregular shapes. Among the vessels are glial cells. H & E, × 90. **Right:** Small and large arteries and veins. Van Gieson and resorcin-fuchsirn, × 90.
nous angioma occurred at the age of 10 years. Until then, the child had exhibited no clinical symptoms, and showed poor scholastic ability in comparison with other children of the same age. The familial tendency to angioma is borne out by the existence of an angioma of the skin in the child’s sister. The oral angioma was treated surgically but unsuccessfully. The cerebral arteriovenous angioma was on the same side as that of the oral cavity, as in the case described by Branch, et al. On the basis of the present case, it would appear that diagnosis of oral venous angioma in early youth may lead to a suspicion of arteriovenous malformation in the brain.

References

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